

## **References**

**Abbott KC, and Agodoa LY, (2002):**

Polycystic kidney disease in patients on the renal transplant waiting list: trends in hematocrit and survival.  
Br Med Clin Nephrol 3: 7.

**Abderrahim E, Hedri H, Laabidi J, et al., (2004):**

Chronic subdural hematoma and autosomal polycystic kidney disease: report of two new cases.  
Nephrology (Carlton) 9: 331-33.

**Adham LM, Soto CE, Inagami T, Cassis L, (2004):**

The intrarenal renin-angiotensin system in autosomal dominant polycystic kidney disease.  
Am J Physiol Renal Physiol 287: F775-88.

**Aguiari G, Banzi M, Gessi S, et al., (2004):**

Deficiency of polycystin-2 reduces  $\text{Ca}^{2+}$  channel activity and cell proliferation in ADPKD lymphoblastoid cells.  
Faseb J 18: 884-86.

**Ahrabi AK, Terryn S, Valenti G, Caron N, Serradeil-Le Gal C, Raufaste D, Nielsen S, Horie S, Verbavatz JM, Devuyst O, (2007):**

PKD1 haplo insufficiency causes a syndrome of inappropriate antidiuresis in mice.  
J Am Soc Nephrol 18: 1740-1753.

**Alehan FK, Gurakan B, Agildere M, (2002):**

Familial arachnoid cysts in association with autosomal dominant polycystic kidney disease.  
Pediatrics 110 (1 part 1): e13.

## **References**

### **Allan EHE and Robert KM, (1992):**

The development of genetics. In: "Elements of Medical Genetics", 8<sup>th</sup> edition, Edited by: Allan E and Robert F, Educational low-priced books Scheme Funded by the British Government ELBS with Churchill-Livingstone, Chapter I, pp 1-10.

### **Alvaro D, Mancino MG, Onori P, et al., (2006):**

Estrogens and the pathophysiology of the biliary tree. World J Gastroenterol 12: 3537-45.

### **Amura CR, Brodsky KS, Groff R, Gattone VH, Voelkel NF, Doctor RB, (2007):**

VEGF receptor inhibition blocks liver cyst growth in pkd2(WS25/ ) mice.

Am J Physiol Cell Physiol 293: C419–C428.

### **Anyatonwu GI, Estrada M, Tian X, Somlo S, Ehrlich BE, (2007):**

Regulation of ryanodine receptor-dependent calcium signaling by polycystin-2.

Proc Natl Acad Sci USA 104: 6454-6459.

### **Arnold HL, and Harrison SA, (2005):**

New advances in evaluation and management of patients with polycystic liver disease.

Am J Gastroenterol 100: 2569-82.

### **Astrinidis A and Henske EP, (2005):**

Tuberous sclerosis complex: linking growth and energy signaling pathways with human disease.

Oncogene 24: 7475-7481.

## References

- =====
- Baba M, Furihata M, Hong SB, Tessarollo L, Haines DC, Southon E, Patel V, Igarashi P, Alvord WG, Leighty R, Yao M, Bernardo M, Ileva L, Choyke P, Warren MB, Zbar B, Linehan WM, Schmidt LS, (2008):**  
Kidney-targeted Birt-Hogg-Dube gene inactivation in a mouse model: Erk1/2 and Akt-mTOR activation, cell hyperproliferation, and polycystic kidneys.  
J Natl Cancer Inst 100: 140–154.
- Badano JL, Teslovich TM, Katsanis N, (2005):**  
The centrosome in human genetic disease.  
Nat Rev Genet 6: 194-205.
- Bae KT, Zhu F, Chapman AB, Torres VE, Grantham JJ, Woodford GLM, Baumgarten DA, King BF Jr, Wetzel LH, Kenney PJ, Brummer ME, Bennett WM, Klahr S, Meyers CM, Zhang X, Thompson PA, Miller JP, (2006):**  
Magnetic resonance imaging evaluation of hepatic cysts in early autosomal dominant polycystic kidney disease: the Consortium for Radiologic Imaging Studies of Polycystic Kidney Disease cohort.  
Clin J Am Soc Nephrol 1: 64-69.
- Bajwa ZH, Gupta S, Warfield CA, Steinman TI, (2001):**  
Pain management in polycystic kidney disease.  
Kidney Int 60: 1631-44.
- Bajwa ZH, Sial KA, Malik AB, Steinman TI, (2004):**  
Pain patterns in patients with polycystic kidney disease.  
Kidney Int 66: 1561-69.

## References

=====

**Banikazemi M, Bultas J, Waldek S, Wilcox WR, Whitley CB, McDonald M, Finkel R, Packman S, Bichet DG, Warnock DG, Desnick RJ, (2007):**

Agalsidase-beta therapy for advanced Fabry disease: A randomized trial.

Ann Intern Med 146: 77–86.

**Barilli A, Visigalli R, Sala R, Gazzola GC, Parolari A, Tremoli E, Bonomini S, Simon A, Closs EI, Dall'Asta V, Bussolati O, (2008):**

In human endothelial cells rapamycin causes mTORC2 inhibition and impairs cell viability and function.

Cardiovasc Res 78: 563–571.

**Basar O, Ibis M, Ucar E, et al., (2006):**

Recurrent pancreatitis in a patient with autosomal dominant polycystic kidney disease.

Pancreatology 6: 160-62.

**Belz MM, Brosnahan FGM, Hughes RL, et al., (2003):**

Recurrence of intracranial aneurysms in autosomal dominant polycystic kidney disease.

Kidney Int 63: 1824-30.

**Berndt TJ, Schiavi S, Kumar R, (2005):**

Phosphatonins and the regulation of phosphorus homeostasis.

Am J Physiol Renal Physiol 289: F1170-82.

**Boca M, Distefano G, Qian F, et al., (2006):**

Polycystin-1 induces resistance to apoptosis through the phosphatidylinositol 3-kinase/Akt signaling pathway.

J Am Soc Nephrol 17: 637-647.

## **References**

**Bonnet CS, Aldred M, von Ruhland C, Harris R, Sandford R, Cheadle JP, (2009):**

Defects in cell polarity underlie TSC and ADPKD-associated cystogenesis.

Hum Mol Genet 18: 2166–2176.

**Brosnahan FGM, Belz MM, McFann KK, Johnson AM, Schrier RW, (2002):**

Relationship between renal volume growth and renal function in autosomal dominant polycystic kidney disease: a longitudinal study.

Am J Kidney Dis 39: 1127-34.

**Brown EM, and Herbert SC, (2009):**

Inherited Diseases of the Calcium-Sensing receptors: Impact on Parathyroid and Renal Function. In: "Genetic Diseases of the Kidney; By Lifton RP, Somlo S, Giebisch GH, Seldin DW", 1<sup>st</sup> edition, Part IID, chapter 15, pp 263-78.

**Bukanov N, Smith LA, Klinger K, Ledbetter SR, Beskrovnaya IO, (2006):**

Long lasting arrest of murine polycystic kidney disease with CDK inhibitor roscovitine.

Nature 444: 949-52.

**Cadnapaphornchai MA, Brosnahan FGM, Duley I, et al., (2005):**

Design and baseline characteristics of participants in the study of antihypertensive therapy in children and adolescents with autosomal dominant polycystic kidney disease (ADPKD).

Contemp Clin Trials 26: 211-22.

## **References**

**Carracedo A, Ma L, Teruya-Feldstein J, Rojo F, Salmena L, Alimonti A, Egia A, Sasaki AT, Thomas G, Kozma SC, Papa A, Nardella C, Cantley LC, Baselga J, Pandolfi PP, (2008):**

Inhibition of mTORC1 leads to MAPK pathway activation through a PI3K-dependent feedback loop in human cancer.

J Clin Invest 118: 3065–3074.

**Casas JP, Chua W, Loukogeorgakis S, Vallance P, Smeeth L, Hingorani AD, MacAllister RJ, (2005):**

Effect of inhibitors of the renin-angiotensin system and other antihypertensive drugs on renal outcomes: systematic review and meta-analysis.

Lancet 366: 2026-2033.

**Chang MY, Parker E, Ibrahim S, et al., (2006):**

Haploinsufficiency of PKD2 is associated with increased tubular cell proliferation and interstitial fibrosis in two murine PKD2 models.

Nephrol Dial Transplant 21: 2078-84.

**Chapman AB, (2008):**

Approaches to Testing New Treatments in Autosomal Dominant Polycystic Kidney Disease: Insights from the CRISP and HALT-PKD Studies.

Clin J Am Soc Nephrol 3: 1197-1204.

**Chapman AB, Grantham JJ, Torres VE, Woodford GLM, Braun W, Oskoui RF, Kelleher C, Schrier RW, Perrone R, Steinman TI, Miskulin D, Meyers CM, Bost JE, Miller JP, Bae KT, (2007):**

## References

- Absence of nephrogenic systemic fibrosis (NSF) following gadolinium exposure in ADPKD individuals with stable CKD.  
J Am Soc Nephrol 18: 837A.
- Chapman AB, Guay-Woodford LM, Grantham JJ, et al., (2003):**  
Renal structure in early autosomal dominant polycystic kidney disease (ADPKD): the Consortium for Radiologic Imaging Studies of Polycystic Kidney Disease (CRISP) cohort.  
Kidney Int 64: 1035-45.
- Chapman AB, Torres VE, Grantham JJ, Shoaf SS, Ouyang JJ, Czerwiec FS, (2005):**  
A phase IIB pilot study of the safety and efficacy of tolvaptan, a vasopressin V<sub>2</sub> receptor antagonist (V2RA) in patients with ADPKD.  
J Am Soc Nephrol 16: 68A.
- Chapuis O, Sockeel P, Pallas G, Pons F, Jancovici R, (2004):**  
Thoracoscopic renal denervation for intractable autosomal dominant polycystic kidney disease–related pain.  
Am J Kidney Dis 43: 161-63.
- Chauveau D, Fakhouri F, Grunfeld JP, (2000):**  
Liver involvement in autosomal dominant polycystic kidney disease: therapeutic dilemma.  
J Am Soc Nephrol 11: 1767-75.
- Chauvet V, Tian X, Husson H, et al., (2004):**  
Mechanical stimuli induce cleavage and nuclear translocation of the polycystin-1 C terminus.  
J Clin Invest 114: 1433-43.

## **References**

**Chobanian AV, Bakris GL, Black HR, Cushman WC, Green LA, Izzo JL Jr, Jones DW, Materson BJ, Oparil S, Wright JT Jr, Roccella EJ, (2003):**

The Seventh Report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure: the JNC 7 report.

J A M A 289: 2560-2572.

**Choo AY, Yoon SO, Kim SG, Roux PP, Blenis J, (2008):**

Rapamycin differentially inhibits S6Ks and 4E-BP1 to mediate cell-type-specific repression of mRNA translation.

Proc Natl Acad Sci USA 105: 17414–17419.

**Clausen P, Feldt-Rasmussen B, Iversen J, Lange M, Eidemak I, Strandgaard S, (2006):**

Flow-associated dilatory capacity of the brachial artery is intact in early autosomal dominant polycystic kidney disease.

Am J Nephrol 26: 335-39.

**Dalgaard OZ, (1957):**

Bilateral polycystic disease of the kidneys: a follow-up of two hundred and eighty-four patients and their families.

Acta Med Scand 328(suppl): 1-255.

**Davila S, Furu L, Gharavi AG, et al., (2004):**

Mutations in SEC63 cause autosomal dominant polycystic liver disease.

Nat Genet 36: 575-77.

**De Rycke M, Georgiou I, Sermon K, et al., (2005):**

PGD for autosomal dominant polycystic kidney disease type 1.

Mol Hum Reprod 11: 65-71.



## References

=====

**Distefano G, Boca M, Rowe I, Wodarczyk C, Ma L, Piontek KB, Germino GG, Pandolfi PP, Boletta A, (2009):**

Polycystin-1 regulates extracellular signal-regulated kinase-dependent phosphorylation of tuberlin to control cell size through mTOR and its downstream effectors S6K and 4EBP1.

Mol Cell Biol 29: 2359–2371.

**Doerken M, Kotsis F, Kottgen M, Braeg S, Walz G, Kuehn W, (2007):**

Suppression of ciliary flow sensing through inducible knock-down of the PKD2 interactor TRPV4.

J Am Soc Nephrol 18: 13A.

**Doulton TW, Malik SAK, He FJ, et al., (2006):**

The effect of sodium and angiotensin-converting enzyme inhibition on the classic circulating renin-angiotensin system in autosomal dominant polycystic kidney disease patients.

J Hypertens 24: 939-45.

**Drenth JP, Te Morsche RH, Smink R, Bonifacino JS, Jansen JB, (2003):**

Germline mutations in PRKCSH are associated with autosomal dominant polycystic liver disease.

Nat Genet 10: 10.

**Dunn MD, Portis AJ, Elbahnasy AM, et al., (2000):**

Laparoscopic nephrectomy in patients with end-stage renal disease and autosomal dominant polycystic kidney disease.

Am J Kidney Dis 35: 720-25.

## **References**

**Ecder T, Chapman A, Brosnahan G, Edelstein C, Johnson A, Schrier R, (2000):**

Effect of antihypertensive therapy on renal function and urinary albumin excretion in hypertensive patients with autosomal dominant polycystic kidney disease.  
Am J Kidney Dis 35: 427-32.

**Ecder T, Edelstein C, Brosnahan FG, et al., (2001):**

Diuretics versus angiotensin-converting enzyme inhibitors in autosomal dominant polycystic kidney disease.  
Am J Nephrol 21: 98-103.

**Everson GT, and Taylor MR, (2005):**

Management of polycystic liver disease.  
Curr Gastroenterol Rep 7: 19-25.

**Fabris L, Cadamuro M, Fiorotto R, et al., (2006):**

Effects of angiogenic factor over-expression by human and rodent cholangiocytes in polycystic liver diseases.  
Hepatology 43: 1001-12.

**Fain PR, McFann KK, Taylor MR, et al., (2005):**

Modifier genes play a significant role in the phenotypic expression of PKD1.  
Kidney Int 67: 1256-67.

**Faivre S, Kroemer G, Raymond E, (2006):**

Current development of mTOR inhibitors as anticancer agents.  
Nat Rev Drug Discov 5: 671–688.

## References

### **Fine LG and Norman JT, (2008):**

Chronic hypoxia as a mechanism of progression of chronic kidney diseases: From hypothesis to novel therapeutics.

Kidney Int 74: 867–872.

### **Fischer DC, Jacoby U, Pape L, Ward CJ, Kuwertz-Broeking E, Renken C, Nizze H, Querfeld U, Rudolph B, Mueller-Wiefel DE, Bergmann C, Haffner D, (2009):**

Activation of the AKT/mTOR pathway in autosomal recessive polycystic kidney disease (ARPKD).

Nephrol Dial Transplant 24: 1819–1827.

### **Foggensteiner L, Bevan AP, Thomas R, et al., (2000):**

Cellular and subcellular distribution of polycystin-2, the protein product of the PKD2 gene.

J Am Soc Nephrol 11: 814-27.

### **Fujishita T, Aoki K, Lane HA, Aoki M, Taketo MM, (2008):**

Inhibition of the mTORC1 pathway suppresses intestinal polyp formation and reduces mortality in Apc Delta 716 mice.

Proc Natl Acad SciUSA 105: 13544–13549.

### **Gao Z, Joseph E, Ruden DM, Lu X, (2004):**

Drosophila PKD2 is haploid-insufficient for mediating optimal smooth muscle contractility.

J Biol Chem 279: 14225-31.

### **Gattone VH, Kinne Q, Torres VE, (2005):**

Efficacy of OPC-41061 in the treatment of murine nephronophthisis.

J Am Soc Nephrol 16: 138A.

## References

=====

**Gattone VH, Sinderson RM, Hornberger TA, Robling AG, (2009):**

Late progression of renal pathology and cyst enlargement is reduced by rapamycin in a mouse model of nephronophthisis.

Kidney Int 76: 178–182.

**Gattone VH, Wang X, Harris PC, Torres VE, (2003):**

Inhibition of renal cystic disease development and progression by a vasopressin V<sub>2</sub> receptor antagonist.

Nat Med 9: 1323-26.

**Gheorghiade M, Gattis WA, O'Connor CM, et al., (2004):**

Effects of tolvaptan, a vasopressin antagonist, in patients hospitalized with worsening heart failure: a randomized controlled trial.

JAMA 291: 1963-1971.

**Gheorghiade M, Konstam MA, Burnett JC Jr, Grinfeld L, Maggioni AP, Swedberg K, Udelson JE, Zannad F, Cook T, Ouyang J, Zimmer C, Orlandi C, (2007):**

Short-term clinical effects of tolvaptan, an oral vasopressin antagonist, in patients hospitalized for heart failure: The EVEREST Clinical Status Trials.

JAMA 297: 1332-1343.

**Gheorghiade M, Niazi I, Ouyang J, et al., (2003):**

Vasopressin V<sub>2</sub>-receptor blockade with tolvaptan in patients with chronic heart failure: results from a double-blind, randomized trial.

Circulation 107: 2690-2696.

## **References**

**Gibbs GF, Huston J, Qian Q, et al., (2004):**

Follow-up of intracranial aneurysms in autosomal dominant polycystic kidney disease.

Kidney Int 65: 1621-27.

**Grampsas SA, Chandhoke PS, Fan J, et al., (2000):**

Anatomic and metabolic risk factors for nephrolithiasis in patients with autosomal dominant polycystic kidney disease.

Am J Kidney Dis 36: 53-57.

**Grantham JJ, (2008):**

Autosomal dominant polycystic kidney disease.

N Engl J Med 359: 1477-85.

**Grantham JJ, Cook LT, Torres VE, Bost JE, Chapman AB, Harris PC, Woodford GLM, Bae KT, (2008):**

Determinants of renal volume in autosomal-dominant polycystic kidney disease.

Kidney Int 73: 108-116.

**Grantham JJ, Torres VE, Chapman AB, Woodford GLM, Bae KT, King BF Jr, Wetzel LH, Baumgarten DA, Kenney PJ, Harris PC, Klahr S, Bennett WM, Hirschman GN, Meyers CM, Zhang X, Zhu F, Miller JP, (2006):**

Volume progression in polycystic kidney disease.

N Engl J Med 354: 2122-2130.

**Gurevich F and Perazella MA, (2009):**

Renal effects of anti-angiogenesis therapy: Update for the internist.

Am J Med 122: 322-328.

## References

**Hanaoka K, and Guggino W, (2000):**

cAMP regulates cell proliferation and cyst formation in autosomal polycystic kidney disease cells.

J Am Soc Nephrol 11: 1179-87.

**Hanaoka K, Qian F, Boletta A, et al., (2000):**

Co-assembly of polycystin-1 and -2 produces unique cation-permeable currents.

Nature 408: 990-94.

**Harris PC, and Torres VE, (2006):**

Understanding pathogenic mechanisms in polycystic kidney disease provides clues for therapy. Curr Opin Nephrol Hypertens 15: 456-63.

**Harris PC, Bae KT, Rossetti S, Torres VE, Grantham JJ, Chapman AB, Woodford GLM, King BF, Wetzel LH, Baumgarten DA, Kenney PJ, Consugar M, Klahr S, Bennett WM, Meyers CM, Zhang QJ, Thompson PA, Zhu F, Miller JP, (2006):**

Cyst number but not the rate of cystic growth is associated with the mutated gene in autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 17: 3013-3019.

**Hasumi H, Baba M, Hong SB, Hasumi Y, Huang Y, Yao M, Valera VA, Linehan WM, Schmidt LS, (2008):**

Identification and characterization of a novel folliculin-interacting protein FNIP2.

Gene 415: 60–67.

**Heinonen PK, Vuento M, Maunola M, Houhala AI, (2002):**

Ovarian manifestations in women with autosomal dominant polycystic kidney disease.

Am J Kidney Dis 40: 504-07.

## **References**

**Hsu YC, Chern JJ, Cai Y, Liu M, Choi KW, (2007):**

Drosophila TCTP is essential for growth and proliferation through regulation of dRheb GTPase.

Nature 445: 785–788.

**Huang J and Manning BD, (2008):**

The TSC1-TSC2 complex: A molecular switchboard controlling cell growth.

Biochem J 412: 179–190.

**Inagawa T, (2001):**

Trends in incidence and case fatality rates of aneurismal subarachnoid hemorrhage in Izumo City, Japan, between 1980-1989 and 1990-1998.

Stroke 32: 1499-507.

**Inoki K, Corradetti MN, Guan KL, (2005):**

Dysregulation of the TSC-mTOR pathway in human disease.

Nat Genet 37: 19-24.

**Inoki K, Ouyang H, Zhu T, Lindvall C, Wang Y, Zhang X, Yang Q, Bennett C, Harada Y, Stankunas K, Wang CY, He X, MacDougald OA, You M, Williams BO, Guan KL, (2006):**

TSC2 integrates Wnt and energy signals via a coordinated phosphorylation by AMPK and GSK3 to regulate cell growth.

Cell 126: 955–968.

**Jafar TH, Stark PC, Schmid CH, et al., (2005):**

The effect of angiotensin-converting-enzyme inhibitors on progression of advanced polycystic kidney disease.

Kidney Int 67: 265-71.

## References

\*\*\*\*\*  
**Jiang ST, Chiou YY, Wang E, et al., (2006):**

Defining a link with autosomal dominant polycystic kidney disease in mice with congenitally low expression of PKD1.

Am J Pathol 168: 205-20.

**Kelleher CL, McFann KK, Johnson AM, Schrier RW, (2004):**

Characteristics of hypertension in young adults with autosomal dominant polycystic kidney disease compared with the general US population.

Am J Hypertens 17 (11 part 1): 1029-34.

**Kielstein R, and Sass HM, (2002):**

Genetics in kidney disease: how much do we want to know.

Am J Kidney Dis 39: 637-52.

**King BF, Reed JE, Bergstralh EJ, Sheedy PF, II, Torres VE, (2000):**

Quantification and longitudinal trends of kidney, renal cyst, and renal parenchyma volumes in autosomal dominant polycystic kidney disease.

J Am Soc Neph 11: 1505-11.

**King BF, Torres VE, Brummer ME, et al., (2003):**

Magnetic resonance measurements of renal blood flow as a marker of disease severity in autosomal-dominant polycystic kidney disease.

Kidney Int 64: 2214-2221.

**Kip SN, Hunter LW, Ren Q, et al., (2005):**

[Ca<sup>2+</sup>]<sup>+</sup> reduction increases cellular proliferation and apoptosis in vascular smooth muscle cells: relevance to the ADPKD phenotype.

Circ Res 96: 873-80.



## References

- =====
- Kistler AD, Poster D, Krauer F, Weishaupt D, Raina S, Senn O, Binet I, Spanaus K, Wuthrich RP, Serra AL, (2009):**  
Increases in kidney volume in autosomal dominant polycystic kidney disease can be detected within 6 months.  
Kidney Int 75: 235–241.
- Klein IH, Ligtenberg G, Oey PL, Koomans HA, Blankestijn PJ, (2001):**  
Sympathetic activity is increased in polycystic kidney disease and is associated with hypertension.  
J Am Soc Nephrol 12: 2427-33.
- Kocaman O, Oflaz H, Yekeler E, et al., (2004):**  
Endothelial dysfunction and increased carotid intima-media thickness in patients with autosomal dominant polycystic kidney disease.  
Am J Kidney Dis 43: 854-60.
- Konstam MA, Gheorghiade M, Burnett JC Jr, Grinfeld L, Maggioni AP, Swedberg K, Udelson JE, Zannad F, Cook T, Ouyang J, Zimmer C, Orlandi C, (2007):**  
Effects of oral tolvaptan in patients hospitalized for worsening heart failure: The EVEREST Outcome Trial.  
J A M A 297: 1319-1331.
- Koulen P, Cai Y, Geng L, et al., (2002):**  
Polycystin-2 is an intracellular calcium release channel.  
Nat Cell Biol 4: 191-97.
- Kumar S, Adeva M, King BF, Kamath PS, Torres VE, (2006):**  
Duodenal diverticulosis in autosomal dominant polycystic kidney disease.  
Nephrol Dial Transplant 21: 3578-78.

## References

**Lederman ED, McCoy G, Conti DJ, Lee EC, (2000):**

Diverticulitis and polycystic kidney disease.

Am Surg 66: 200-03.

**Lee DI, and Clayman RV, (2004):**

Hand-assisted laparoscopic nephrectomy in autosomal dominant polycystic kidney disease.

J Endourol 18: 379-82.

**Lee DI, Andreoni CR, Rehman J, et al., (2003):**

Laparoscopic cyst decortication in autosomal dominant polycystic kidney disease: impact on pain, hypertension, and renal function.

J Endourol 17: 345-54.

**Lee SE, John SH, Lim JH, Rhew JY, (2008):**

Very late stent thrombosis associated with multiple stent fractures and persistent aneurysm formation after sirolimus-eluting stent implantation.

Circ J 72: 1201–1204.

**Li A, Davila S, Furu L, et al., (2003):**

Mutations in PRKCSH cause isolated autosomal dominant polycystic liver disease.

Am J Hum Genet 72: 691-703.

**Li VM, Cianfrone P, Damiano R, Fuiano G, (2003):**

Infertility in adults with polycystic kidney disease.

Nephrol Dial Transplant 18: 190-91.

**Li Y, Wright JM, Qian F, Germino GG, Guggino WB, (2005):**

Polycystin 2 interacts with type I inositol 1,4,5–trisphosphate receptor to modulate intracellular Ca<sup>2+</sup> signaling.

J Biol Chem 280: 41298-306.

## **References**

### **Low SH, Vasanth S, Larson CH, et al., (2006):**

Polycystin-1, STAT6, and P100 function in a pathway that transduces ciliary mechano-sensation and is activated in polycystic kidney disease.

Dev Cell 10: 57-69.

### **Lumiaho A, Ikaheimo R, Miettinen R, et al., (2001):**

Mitral valve prolapse and mitral regurgitation are common in patients with polycystic kidney disease type 1.

Am J Kidney Dis 38: 1208-16.

### **MacKinnon M, Shurraw S, Akbari A, Knoll GA, Jaffey J, Clark HD, (2006):**

Combination therapy with an angiotensin receptor blocker and an ACE inhibitor in proteinuric renal disease: a systematic review of the efficacy and safety data.

Am J Kidney Dis 48: 8-20.

### **Magistroni R, He N, Wang K, et al., (2003):**

Genotype-renal function correlation in type 2 autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 14: 1164-74.

### **Martinez-Lemus LA, Hill MA, Meininger GA, (2009):**

The plastic nature of the vascular wall: a continuum of remodeling events contributing to control of arteriolar diameter and structure.

Physiology (Bethesda) 24: 45–57.

### **McPherson EA, Luo Z, Brown RA, et al., (2004):**

Chymase-like angiotensin II-generating activity in end-stage human autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 15: 493-500.

## **References**

=====

**McPherson EA, Luo Z, Brown RA, Lebard LS, Corless CC, Speth RC, Bagby SP, (2004):**

Chymase-like angiotensin II-generating activity in end-stage human autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 15: 493-500.

**Morel N, Vandenberg G, Ahrabi AK, Caron N, Desjardins F, Balligand JL, Horie S, Devuyst O, (2009):**

PKD1 haploinsufficiency is associated with altered vascular reactivity and abnormal calcium signaling in the mouse aorta.

Pflugers Arch 457: 845–856.

**Mutig K, Paliege A, Kahl T, Jons T, Muller-Esterl WP, Bachmann S, (2007):**

Vasopressin V2 receptor expression along rat, mouse, and human renal epithelia with focus on TAL.

Am J Physiol Renal Physiol 293: F1166-F1177.

**Nagao S, Kazuhiro N, Katsuyama M, Kurahashi H, Marunouchi T, Takahashi H, Wallace DP, (2006):**

Increased water intake decreases progression of polycystic kidney disease in the PCK rat.

J Am Soc Nephrol 17: 228-235.

**Naitoh H, Shoji H, Ishikawa I, et al., (2005):**

Intraductal papillary mucinous tumor of the pancreas associated with autosomal dominant polycystic kidney disease.

J Gastrointest Surg 9: 843-45.

## **References**

**Nauli SM, Alenghat FJ, Luo Y, et al., (2003):**

Polycystins 1 and 2 mediate mechano-sensation in the primary cilium of kidney cells.

Nat Genet 33: 129-37.

**Nichols MT, Gidey E, Matzakos T, et al., (2004):**

Secretion of cytokines and growth factors into autosomal dominant polycystic kidney disease liver cyst fluid.

Hepatology 40: 836-46.

**Nicolau C, Torra R, Bianchi L, et al., (2000):**

Abdominal sonographic study of autosomal dominant polycystic kidney disease.

J Clin Ultrasound 28: 277-82.

**Nishio S, Hatano M, Nagata M, et al., (2005):**

Pkd1 regulates immortalized proliferation of renal tubular epithelial cells through p53 induction and JNK activation.

J Clin Invest 115: 910-918.

**Nutahara K, Higashihara E, Horie S, et al., (2005):**

Calcium channel blocker versus angiotensin II receptor blocker in autosomal dominant polycystic kidney disease.

Nephron Clin Pract 99: c18-23.

**Omori S, Hida M, Fujita H, et al., (2006):**

Extracellular signal-regulated kinase inhibition slows disease progression in mice with polycystic kidney disease.

J Am Soc Nephrol 17: 1604-14.

**Orth SR, (2000):**

Smoking; a renal risk factor.

Nephron 86: 12-26.

## References

**Ortiz PA, (2006):**

cAMP increases surface expression of NKCC2 in rat thick ascending limbs: Role of VAMP.

Am J Physiol Renal Physiol 290: F608-F616.

**Osawa H, Nakamura N, Shirato K, et al., (2006):**

Losartan, an angiotensin-II receptor antagonist, retards the progression of advanced renal insufficiency.

Tohoku J Exp Med 209: 7-13.

**Park JY, Schutzer WE, Lindsley JN, Bagby SP, Oyama TT, Anderson S, Weiss RH, (2007):**

p21 is decreased in polycystic kidney disease and leads to increased epithelial cell cycle progression: roscovitine augments p21 levels.

Br Med Clin Nephrol 8: 12.

**Paterson AD, Magistroni R, He N, et al., (2005):**

Progressive loss of renal function is an age-dependent heritable trait in type 1 autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 16: 755-62.

**Paterson AD, Wang KR, Lupea D, Hyslop StGP, Pei Y, (2002):**

Recurrent fetal loss associated with bilineal inheritance of type 1 autosomal dominant polycystic kidney disease.

Am J Kidney Dis 40: 16-20.

**Pazour GJ, San Agustin JT, Follit JA, Rosenbaum JL, Witman GB, (2002):**

Polycystin-2 localizes to kidney cilia and the ciliary level is elevated in orpk mice with polycystic kidney disease.

Curr Biol 12: R378-80.

## **References**

**Pei Y, Paterson AD, Wang KR, et al., (2001):**

Bilineal disease and trans-heterozygotes in autosomal dominant polycystic kidney disease.

Am J Hum Genet 68: 355-63.

**Perrett GS, Kim K, Ibarra C, et al., (2001):**

Polycystin-2, the protein mutated in autosomal dominant polycystic kidney disease (ADPKD), is a  $\text{Ca}^{2+}$ -permeable non-selective cation channel.

Proc Natl Acad Sci USA 98: 1182-87.

**Persu A, Duyme M, Pirson Y, et al., (2004):**

Comparison between siblings and twins supports a role for modifier genes in ADPKD.

Kidney Int 66: 2132-36.

**Pirson Y, Chauveau D, Torres VE, (2002):**

Management of cerebral aneurysms in autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 13: 269-76.

**Praetorius HA and Spring KR, (2005):**

A physiological view of the primary cilium.

Annu Rev Physiol 67: 515-29.

**Qian F, Boletta A, Bhunia AK, et al., (2002):**

Cleavage of polycystin-1 requires the receptor for egg jelly domain and is disrupted by human autosomal dominant polycystic kidney disease 1-associated mutations.

Proc Natl Acad Sci USA 99: 16981-86.

**Qian Q, Du H, King BF, Kumar S, Dean PG, Cosio FG, Torres VE, (2008):**

Sirolimus reduces polycystic liver volume in ADPKD patients.

## References

- J Am Soc Nephrol 19: 631–638.
- Qian Q, Hunter LW, Du H, Ren Q, Han Y, Sieck GC, (2007):**  
PKD<sub>2</sub><sup>+/-</sup> vascular smooth muscles develop exaggerated vasocontraction in response to phenylephrine stimulation.  
J Am Soc Neph 18: 485-93.
- Qian Q, Hunter LW, Li M, et al., (2003):**  
PKD2 haplo-insufficiency alters intracellular calcium in vascular smooth muscle cells.  
Hum Mol Genet 12: 1875-80.
- Qian Q, Li A, King BF, et al., (2003):**  
Clinical profile of autosomal dominant polycystic liver disease.  
Hepatology 37: 164-171.
- Qian Q, Li M, Cai Y, et al., (2003):**  
Analysis of the polycystins in aortic vascular smooth muscle cells.  
J Am Soc Nephrol 14: 2280-87.
- Reichardt W, Romaker D, Becker A, Buechert M, Walz G, von Elverfeldt D, (2009):**  
Monitoring kidney and renal cyst volumes applying MR approaches on a rapamycin treated mouse model of ADPKD.  
MAGMA 22: 143–149.
- Reis F, Parada B, Teixeira de Lemos, Garrido P, Dias A, Piloto N, Baptista S, Sereno J, Eufra´sio P, Costa E, Rocha-Pereira P, Santos-Silva A, Figueiredo A, Mota A, Teixeira F, (2009):**  
Hypertension induced by immunosuppressive drugs: A comparative analysis between sirolimus and cyclosporine.  
Transplant Proc 41: 868–873.



## **References**

### **Renal Data System US USRDS, (1999):**

Annual data report. Bethesda: National Institutes of Health, 1999.

### **Reuss BE, Holubec K, Rajaraman S, (2001):**

Angiogenesis in autosomal dominant polycystic kidney disease.

Kidney Int 60: 37-45.

### **Reynolds DM, Falk CT, Li AR, et al., (2000):**

Identification of a locus for autosomal dominant polycystic liver disease, on chromosome 19p13.2-13.1.

Am J Hum Genet 67: 1598-604.

### **Rossetti S, Burton S, Strmecki L, et al., (2002):**

The position of the polycystic kidney disease 1 (PKD1) gene mutation correlates with the severity of renal disease.

J Am Soc Nephrol 13: 1230-37.

### **Rossetti S, Chauveau D, Kubly V, et al., (2003):**

Association of mutation position in polycystic kidney disease 1 (PKD1) gene and development of a vascular phenotype.

Lancet 361: 2196-201.

### **Rossetti S, Chauveau D, Walker D, et al., (2002):**

A complete mutation screen of the ADPKD genes by DHPLC.

Kidney Int 61: 1588-99.

### **Rossetti S, Strmecki L, Gamble V, et al., (2001):**

Mutation analysis of the entire PKD1 gene: Genetic and diagnostic implications.

Am J Hum Genet 68: 46-63.

## **References**

**Rovers BCP, de Sevaux RG, van Hamersvelt HW, Corstens FH, Oyen WJ, (2003):**

Diagnosis of renal and hepatic cyst infections by 18-F-fl uorodeoxyglucose positron emission tomography in autosomal dominant polycystic kidney disease.

Am J Kidney Dis 41: E18-21.

**Rozanski J, Kozłowska I, Myslak M, et al., (2005):**

Pretransplant nephrectomy in patients with autosomal dominant polycystic kidney disease.

Transplant Proc 37: 666-68.

**Ruggenenti P, Remuzzi A, Odeh P, et al., (2005):**

Safety and efficacy of long-acting somatostatin treatment in autosomal dominant polycystic kidney disease.

Kidney Int 68: 206-16.

**Rundle DR, Gorbsky G, Tsiokas L, (2004):**

PKD2 interacts and co-localizes with mDial1 to mitotic spindles of dividing cells.

J Biol Chem 279: 29728-39.

**Sakurai Y, Shoji M, Matsubara T, et al., (2001):**

Pancreatic ductal adenocarcinoma associated with Potter type III cystic disease.

J Gastroenterol 36: 422-28.

**Sarbassov DD, Ali SM, Kim DH, Guertin DA, Latek RR, Bromage EH, Tempst P, Sabatini DM, (2004):**

Rictor, a novel binding partner of mTOR, defines a rapamycin-insensitive and raptor-independent pathway that regulates the cytoskeleton.

Curr Biol 14: 1296–1302.

## References

- =====
- Sarbassov DD, Ali SM, Sengupta S, Sheen JH, Hsu PP, Bagley AF, Markhard AL, Sabatini DM, (2006):**  
Prolonged rapamycin treatment inhibits mTORC2 assembly and Akt/PKB.  
Mol Cell 22: 159–168.
- Sarnak MJ, Greene T, Wang X, et al., (2005):**  
The effect of a lower target blood pressure on the progression of kidney disease: long-term follow-up of the modification of diet in renal disease study.  
Ann Intern Med 142: 342-51.
- Sasaki M, Katayanagi K, Watanabe K, Takasawa K, Nakanuma Y, (2002):**  
Intrahepatic cholangio-carcinoma arising in autosomal dominant polycystic kidney disease.  
Virchows Arch 441: 98-100.
- Sauter D, Fernandes S, Goncalves-Mendes N, Boulkroun S, Bankir L, Loffing J, Bouby N, (2006):**  
Long-term effects of vasopressin on the subcellular localization of ENaC in the renal collecting system.  
Kidney Int 69: 1024-1032.
- Scheffers M, van der Bent P, Prins F, et al., (2000):**  
Subcellular localization of polycystin-1 during adhesion complex assembly.  
The Netherlands PKD Workshop, 53.
- Schrier RW, Belz MM, Johnson AM, et al., (2004):**  
Repeat imaging for intracranial aneurysms in patients with autosomal dominant polycystic kidney disease with initially negative studies: a prospective ten-year follow-up.  
J Am Soc Nephrol 15: 1023-28.

## References

\*\*\*\*\*  
**Schrier RW, Gross P, Gheorghiade M, Berl T, Verbalis JG, Czerwiec FS, Orlandi C, (2006):**

Tolvaptan, a selective oral vasopressin V2-receptor antagonist, for hyponatremia.

N Engl J Med 355: 2099-2112.

**Seeman T, Dusek J, Vondrak K, Blahova K, Simkova E, Kreisinger J, Dvorak P, Kyncl M, Hribal Z, Janda J, (2004):**

Renal concentrating capacity is linked to blood pressure in children with autosomal dominant polycystic kidney disease.

Physiol Res 53: 629-634.

**Seeman T, Dusek J, Vondrichova H, et al., (2003):**

Ambulatory blood pressure correlates with renal volume and number of renal cysts in children with autosomal dominant polycystic kidney disease.

Blood Press Monit 8: 107-10.

**Serra AL, Kistler AD, Poster D, Strucker M, Wuthrich RP, Weishaupt D, Tschirch F, (2007):**

Clinical proof-of-concept trial to assess the therapeutic effect of sirolimus in patients with autosomal dominant polycystic kidney disease: SUISSE ADPKD study.

Br Med Clin Nephrol 8: 13.

**Serra V, Markman B, Scaltriti M, Eichhorn PJ, Valero V, Guzman M, Botero ML, Llonch E, Atzori F, Di CS, Maira M, Garcia-Echeverria C, Parra JL, Arribas J, Baselga J, (2008):**

NVP-BEZ235, a dual PI3K/mTOR inhibitor, prevents PI3K signaling and inhibits the growth of cancer cells with activating PI3K mutations.

Cancer Res 68: 8022–8030.

## **References**

**Shah OJ and Hunter T, (2006):**

Turnover of the active fraction of IRS1 involves raptor-mTOR- and S6K1-dependent serine phosphorylation in cell culture models of tuberous sclerosis.

Mol Cell Biol 26: 6425–6434.

**Shillingford JM, Murcia NS, Larson CH, Low SH, Hedgepeth R, Brown N, Flask CA, Novick AC, Goldfarb DA, Kramer-Zucker A, Walz G, Piontek KB, Germino GG, Weimbs T, (2006):**

The mTOR pathway is regulated by polycystin-1, and its inhibition reverses renal cystogenesis in polycystic kidney disease.

Proc Natl Acad Sci USA 103: 5466-5471.

**Shillingford JM, Piontek KB, Germino GG, Weimbs T, (2010):**

Rapamycin ameliorates PKD resulting from conditional inactivation of pkd1.

J Am Soc Nephrol 21: 489–497.

**Silberberg M, Charron AJ, Bacallao R, Ness WA, (2005):**

Mispolarization of desmosomal proteins and altered intercellular adhesion in autosomal dominant polycystic kidney disease.

Am J Physiol Renal Physiol 288: F1153-63.

**Sinha SS, Pham MX, Vagelos RH, Perlroth MG, Hunt SA, Lee DP, Valantine HA, Yeung AC, Fearon WF, (2008):**

Effect of rapamycin therapy on coronary artery physiology early after cardiac transplantation.

Am Heart J 155: 889.e1–e6.

## References

**Sise C, Kusaka M, Wetzel LH, et al., (2000):**

Volumetric determination of progression in autosomal dominant polycystic kidney disease by computed tomography.

Kidney Int 58: 2492-501.

**Soliman AR, Ismail E, Zamil S, Lotfy A, (2009):**

Sirolimus therapy for patients with adult polycystic kidney disease: A pilot study.

Transplant Proc 41: 3639–3641.

**Spirli C, Okolicsanyi S, Fiorotto R, Fabris L, Cadamuro M, Lecchi S, Tian X, Somlo S, Strazzabosco M, (2009):**

Mammalian target of rapamycin regulates vascular endothelial growth factor-dependent liver cyst growth in polycystin-2-defective mice.

Hepatology 51: 1–11.

**Stengel B, Billon S, Van Dijk PC, et al., (2003):**

Trends in the incidence of renal replacement therapy for end-stage renal disease in Europe, 1990-1999.

Nephrol Dial Transplant 18: 1824-33.

**Strippoli GF, Craig M, Deeks JJ, Schena FP, Craig JC, (2004):**

Effects of angiotensin converting enzyme inhibitors and angiotensin II receptor antagonists on mortality and renal outcomes in diabetic nephropathy: systematic review.

Br Med J 329: 828.

**Sweeney WE, Chen Y, Nakanish K, Frost P, Avner ED, (2000):**

Treatment of polycystic kidney disease with a novel tyrosine kinase inhibitor.

Kidney Int 57: 33-40.

## References

=====

**Sweeney WE Jr, and Avner ED, (2006):**

Molecular and cellular pathophysiology of autosomal recessive polycystic kidney disease (ARPKD).

Cell Tissue Res 326: 671-85.

**Tahvanainen P, Tahvanainen E, Reijonen H, Halme L, Kaariainen H, Hockerstedt K, (2003):**

Polycystic liver disease is genetically heterogeneous: clinical and linkage studies in eight Finnish families.

J Hepatol 38: 39-43.

**Tao Y, Kim J, Schrier RW, Edelstein CL, (2005):**

Rapamycin markedly slows disease progression in a rat model of polycystic kidney disease.

J Am Soc Nephrol 16: 46-51.

**Tao Y, Kim J, Yin Y, Zafar I, Falk S, He Z, Faubel S, Schrier RW, Edelstein CL, (2007):**

VEGF receptor inhibition slows the progression of polycystic kidney disease.

Kidney Int 72: 1358–1366.

**Tjio JH, and Levan A, (1956):**

The chromosome number of man.

Hereditas 42: 1-6. (Quoted from Alan & Robert, 1992).

**Torres VE, (2008):**

Role of Vasopressin Antagonists.

Clin J Am Soc Nephrol 3: 1212–1218.

**Torres VE, (2006):**

Water for ADPKD? Probably, yes.

J Am Soc Nephrol 17: 2089-91.

**Torres VE, and Grantham J, (2008):**

Cystic diseases of the kidney. In: "Brenner B, The Kidney", Saunders-Elsevier, Philadelphia, Vol 6, pp 1428-62.

## **References**

### **Torres VE, and Harris PC, (2006):**

Mechanisms of disease: autosomal dominant and recessive polycystic kidney diseases.

Nat Clin Prac Nephro 2: 40-54.

### **Torres VE, Cai Y, Chen X, et al., (2001):**

Vascular expression of polycystin 2.

J Am Soc Nephrol 12: 1-9.

### **Torres VE, Grantham JJ, Chapman AB, Watnick T, Kedzierski K, Ouyang J, Orlandi C, Czerwiec FS, Investigators T, (2007):**

Phase 2 open-label study to determine safety, tolerability and efficacy of split-dose tolvaptan in ADPKD.

J Am Soc Nephrol 18: 361-362A.

### **Torres VE, Harris PC, Pirson Y, (2007):**

Autosomal dominant polycystic kidney disease.

Lancet 369: 1287-301.

### **Torres VE, King BF, Chapman AB, Brummer ME, Bae KT, Glockner JF, Arya K, Risk D, Felmlee JP, Grantham JJ, Woodford GLM, Bennett WM, Klahr S, Meyers CM, Zhang X, Thompson PA, Miller JP, (2007):**

Magnetic resonance measurements of renal blood flow and disease progression in autosomal dominant polycystic kidney disease.

Clin J Am Soc Nephrol 2: 112–120.

### **Torres VE, Sweeney WE Jr, Wang X, et al., (2004):**

Epidermal growth factor receptor tyrosine kinase inhibition is not protective in PCK rats.

Kidney Int 66: 1766-1773.



## References

=====

**Torres VE, Wang X, Qian Q, Somlo S, Harris PC, Gattone VH, (2004):**

Effective treatment of an orthologous model of autosomal dominant polycystic kidney disease.

Nat Med 10: 363-64.

**Torres VE, Wang X, Ward CJ, Grantham JJ, Chapman AB, Ouyang J, Shoaf SE, Czerwiec FS, (2005):**

Urine aquaporin 2 and cyclic AMP responses to tolvaptan administration in autosomal dominant polycystic kidney disease.

J Am Soc Nephrol 16: 361A.

**Turkmen K, Oflaz H, Uslu B, Cimen AO, Elitok A, Kasikcioglu E, Alisir S, Tufan F, Namli S, Uysal M, Ecder T, (2008):**

Coronary flow velocity reserve and carotid intima media thickness in patients with autosomal dominant polycystic kidney disease: From impaired tubules to impaired carotid and coronary arteries.

Clin J Am Soc Nephrol 3: 986–991.

**Valente JF, (2001):**

Laparoscopic renal denervation for intractable ADPKD-related pain.

Nephrol Dial Transplant 16: 160.

**van Dijk MA, Breuning MH, Duiser R, van Es LA, Westendorp RG, (2003):**

No effect of enalapril on progression in autosomal dominant polycystic kidney disease.

Nephrol Dial Transplant 18: 2314-20.

=====

## References

\*\*\*\*\*

**van Leeuwen LIS, Dauwerse JG, Baelde HJ, et al., (2004):**

Lowering of PKD1 expression is sufficient to cause polycystic kidney disease.

Hum Mol Genet 13: 3069-77.

**Vassilev PM, Guo L, Chen XZ, et al., (2001):**

Polycystin-2 is a novel cation channel implicated in defective intracellular  $\text{Ca}^{2+}$  homeostasis in polycystic kidney disease.

Biochem Biophys Res Commun 282: 341-50.

**Wahl PR, Le Hir M, Voetseder A, Arcaro A, Starke A,**

**Waeckerle-Men Y, Serra AL, Wuthrich RP, (2007):**

Mitotic activation of Akt signaling pathway in Han:SPRD rats with polycystic kidney disease.

Nephrology (Carlton) 12: 357-363.

**Wahl PR, Serra AL, Le Hir M, Molle KD, Hall MN,**

**Wuthrich RP, (2006):**

Inhibition of mTOR with sirolimus slows disease progression in Han: SPRD rats with autosomal dominant polycystic kidney disease (ADPKD).

Nephrol Dial Transplant 21: 598-604.

**Wakai K, Nakai S, Kikuchi K, et al., (2004):**

Trends in incidence of end-stage renal disease in Japan, 1983-2000: age-adjusted and age-specific rates by gender and cause.

Nephrol Dial Transplant 19: 2044-52.

**Walz G, (2006):**

Therapeutic approaches in autosomal dominant polycystic kidney disease (ADPKD): is there light at the end of the tunnel?

Nephrol Dial Transplant 21: 1752-57.

\*\*\*\*\*

## References

=====

**Wang D, Iversen J, Wilcox CS, Strandgaard S, (2003):**

Endothelial dysfunction and reduced nitric oxide in resistance arteries in autosomal dominant polycystic kidney disease.

Kidney Int 64: 1381-88.

**Wang S, Gattone VH, Somlo S, Harris PC, Torres VE, (2005):**

Effectiveness of OPC-41061 on polycystic kidney disease development in Pkd2WS25.

J Am Soc Nephrol 16: 361A.

**Wang X, Gattone IIVH, Harris PC, Torres VE, (2005):**

Effectiveness of vasopressin V<sub>2</sub> receptor antagonists OPC-31260 and OPC-41061 on polycystic kidney disease development in the PCK rat.

J Am Soc Nephrol 16: 846-51.

**Wang X, Wu Y, Ward CJ, Harris PC, Torres VE, (2008):**

Vasopressin directly regulates cyst growth in the PCK rat.

J Am Soc Nephrol 19: 102-108.

**Weimbs T, (2007):**

Polycystic kidney disease and renal injury repair: Common pathways, fluid flow, and the function of polycystin-1.

Am J Physiol Renal Physiol 293: F1423–F1432.

**Wiebers DO, Whisnant JP, Huston J, et al., (2003):**

Unruptured intracranial aneurysms: natural history, clinical outcome, and risks of surgical and endovascular treatment.

Lancet 362: 103-10.

=====

## References

=====

**Wijedicks EF, Torres VE, Schievink WI, (2000):**

Chronic subdural hematoma in autosomal dominant polycystic kidney disease.

Am J Kidney Dis 35: 40-43.

**Wilson PD, (2004):**

Polycystic kidney disease.

N Engl J Med 350: 151-164.

**Wilson SJ, Amsler K, Hyink DP, et al., (2006):**

Inhibition of HER-2(neu/ErbB2) restores normal function and structure to polycystic kidney disease (PKD) epithelia.

Biochim Biophys Acta 1762: 647-55.

**Wolf G and Ritz E, (2005):**

Combination therapy with ACE inhibitors and angiotensin II receptor blockers to halt progression of chronic renal disease: pathophysiology and indications.

Kidney Int 67: 799-812.

**Woodward S and Leppert M, (1986):**

A closely linked genetic marker for cystic fibrosis.

Nature 318: 382-384.

**Wu G, Tian X, Nishimura S, et al., (2002):**

Trans-heterozygous PKD1 and PKD2 mutations modify expression of polycystic kidney disease.

Hum Mol Genet 11: 1845-54.

**Wu M, Wahl PR, Le HM, Wackerle-Men Y, Wuthrich RP, Serra AL, (2007):**

Everolimus retards cyst growth and preserves kidney function in a rodent model for polycystic kidney disease.

Kidney Blood Press Res 30: 253–259.

## References

=====

**Wu Y, Dai XQ, Li Q, Chen CX, Mai W, Hussain Z, Long W, Montalbetti N, Li G, Glynne R, Wang S, Cantiello HF, Wu G, Chen XZ, (2006):**

Kinesin-2 mediates physical and functional interactions between polycystin-2 and fibrocystin.

Hum Mol Genet 15: 3280-3292.

**Wullschleger S, Loewith R, Hall MN, (2006):**

TOR signaling in growth and metabolism.

Cell 124: 471-484.

**Xue Q, Nagy JA, Manseau EJ, Phung TL, Dvorak HF, Benjamin LE, (2009):**

Rapamycin inhibition of the Akt/mTOR pathway blocks select stages of VEGF-A164-driven angiogenesis, in part by blocking S6Kinase.

Arterioscler Thromb Vasc Biol 29: 1172–1178.

**Yamaguchi T, Hempson SJ, Reif GA, Hedge AM, Wallace DP, (2006):**

Calcium restores a normal proliferation phenotype in human polycystic kidney disease epithelial cells.

J Am Soc Nephrol 17: 178-87.

**Yamaguchi T, Nagao S, Wallace DP, et al., (2003):**

Cyclic AMP activates B-Raf and ERK in cyst epithelial cells from autosomal dominant polycystic kidneys.

Kidney Int 63: 1983-94.

**Yamaguchi T, Wallace DP, Magenheimer BS, Hempson SJ, Grantham JJ, Calvet JP, (2004):**

Calcium restriction allows cAMP activation of the B-Raf/ERK pathway, switching cells to a cAMP-dependent growth-stimulated phenotype.

J Biol Chem 279: 40419-30.

## References

=====

**Yang J, Zhang S, Zhou Q, Guo H, Zhang K, Zheng R, Xiao C, (2007):**

PKHD1 gene silencing may cause cell abnormal proliferation through modulation of intracellular calcium in autosomal recessive polycystic kidney disease.

J Biochem Mol Biol 40: 467-474.

**Yoder BK, Hou X, Woodford GLM, (2002):**

The polycystic kidney disease proteins, polycystin-1, polycystin-2, polaris, and cystin, are co-localized in renal cilia.

J Am Soc Nephrol 13: 2508-16.

**Zafar I, Belibi FA, He Z, Edelstein CL, (2009):**

Long-term rapamycin therapy in the Han:SPRD rat model of polycystic kidney disease (PKD).

Nephrol Dial Transplant 24: 2349–2353.

**Zeng LH, Xu L, Gutmann DH, Wong M, (2008):**

Rapamycin prevents epilepsy in a mouse model of tuberous sclerosis complex.

Ann Neurol 63: 444–453.

**Zhang H, Bajraszewski N, Wu E, Wang H, Moseman AP, Dabora SL, Griffin JD, Kwiatkowski DJ, (2007):**

PDGFRs are critical for PI3K/Akt activation and negatively regulated by mTOR.

J Clin Invest 117: 730–738.

**Zhang T, Wang L, Xiong X, Mao Z, Wang L, Mei C, (2009):**

Mycophenol at emofetil versus rapamycinin Han: SPRD rats with polycystic kidney disease.

Biol Res 42: 437–444.