Summer 8-3-2017

Polycystic Kidney Disease

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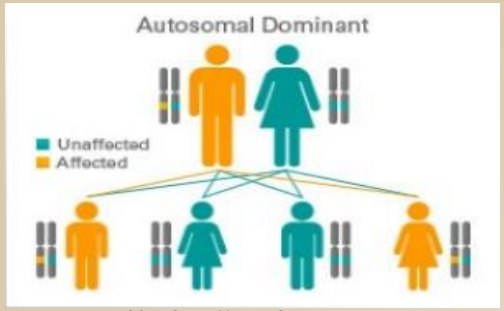

Image retrieved from https://www.physio-pedia.com/Polycystic_Kidney_Disease

INTRODUCTION

This nurse is currently employed at an outpatient ambulatory hemodialysis (HD) clinic. National statistics revealed more than 660,000 Americans being treated for end stage renal disease (ESRD) and at the state level, Ohio has 16,182 individuals which are on routine HD with the primary causes attributed to diabetes and hypertension (HTN) (National Kidney Foundation [NKF], 2017). This nurse discovered a small patient population ascertained ESRD from an inherited familial disorder, particularly polycystic kidney disease (PKD). PKD is the fourth leading cause kidney failure and approximately 600,000 individuals have been diagnosed with PKD in the United States (U.S.) (NKF, 2017). PKD causes multiple fluid cysts to grow in the kidneys, depending on the size and amount they can damage the kidney, decrease kidney function, and lead to renal failure (NKF, 2017).

There are three types of PKD: autosomal dominant polycystic kidney disease (ADPKD), autosomal recessive polycystic kidney disease (ARPKD), and acquired cystic kidney disease (ACKD). ADPKD, which is the most predominant occurring in 300,000 to 600,000 individuals per year (Srivastava & Patel, 2014). ADPKD is the most commonly inherited human renal disease with mutations on two genes, PKD1 and PKD2, mutations from PKD1 account for 85% of ADPKD cases (Sweeney Jr & Avner, 2014). The PKD1 gene is located on chromosome 16 and the PKD2 gene is located on chromosome 4, both genes encode for membrane proteins polycystin-1 and polycystin-2, mutations lead to increased levels of cyclic adenosine monophosphate causing cystogenesis (Srivastava & Patel, 2014). Autosomal dominant diseases affect males and females equally, 50% of the offspring of affected parent will have the disease, and generations are not skipped. Most patients with PKD do not present with clinical manifestations until later in in life due to the slow growth of the cysts which contributes to a loss of nephrons and a decline in the glomerular filtration rate (GFR) (Srivastava & Patel, 2014).

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SIGNS & SYMPTOMS

Renal Cysts

•Multiple fluid filled cysts form within the kidneys from focal proliferation of single tubular epithelial cells.

Hypertension (HTN)

•A risk factor associated with a decline in renal function, development of left ventricular hypertrophy, and increased morbidity.
•The most common manifestation of ADPKD and occurs in 50-70% of cases (Chebib & Torres, 2016).

Pain

•Pain can be present in back, chest, abdominal, or flank.
•Pain is reported in 60% of the cases of adults with ADPKD (Chebib & Torres, 2016).

Urinary Tract Infections

•More common with women and are caused by gram negative bacteria.
•Occurs in 30-50% of the patients (Chebib & Torres, 2016).

Nephrolithiasis

•Renal stones usually composed of uric acid and/or calcium oxalate. Twice as common in ADKPD.
•Patients with ADPKD patients have a decrease in urine citrate, a substance which prevents the formation
•of kidney stones.

Hematuria

UNDERLYING PATHOPHYSIOLOGY

- Polycystins are proteins which regulate intracellular calcium signaling and are found in tissues such as renal tubular epithelia, hepatic bile ducts, and pancreatic bile ducts (Harris &. Torres, 2014).
- Mutations in PKD1 or PKD2 causes a reduction in intracellular calcium, an increase in cyclic adenosine monophosphate (cAMP), activation of protein kinase A, and an increase in sensitivity of collecting duct principal cells to the constant tonic effect of vasopressin.
- A decrease in calcium signaling and an increase in cAMP causes impaired tubulogenesis. cell proliferation, increased fluid secretion, and interstitial inflammation (Silverman, Desai, & Lerma, 2015).

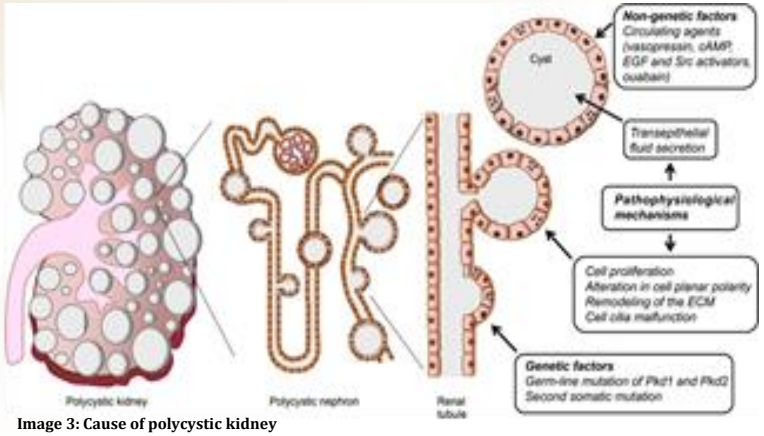

Image 3: Cause of polycystic kidney
Image retrieved from http://healthsaline.com/polycystic-kidney-disease.html

SIGNIFICANCE OF PATHOPHYSIOLOGY

- Cyst development occurs from dilatations in intact tubules which are connected to nephrons.
- Increased cell proliferation and fluid secretion lead to cyst growth causing displacement and loss of the normal renal parenchyma.
- Increased cAMP decreases intracellular calcium signaling increasing the size of cysts by increasing fluid secretion.
- Impairs renal function and leads to ESRD (Cronec-Le Gall et al., 2013).
- Renal cysts compress the renal vasculature resulting in intrarenal ischemia activating the renin-angiotensin-aldosterone system (RAAS).
- Chronic pain caused by cysts rupturing or enlarged kidney compressing other structures (Ong, Devuyst, Knebelmann, & Walz, 2015).
- Acute pain is associated with kidney cyst hemorrhage, infection, or stones.
- Urinary tract infections (UTI) are caused by infected renal cysts.
- Cysts block the renal tubules preventing normal drainage causing crystals to form and creating kidney stones.
- Hematuria is caused by the rupturing of cysts or of the small blood vessels around the cysts (Silverman, Desai, & Lerma, 2015).

IMPLICATIONS FOR NURSING CARE

•Take patient history and perform assessment.

•Monitor vital signs, particularly blood pressure.

•Palpate bilaterally for flank masses (Yrad & Humphreys, 2017).

•Monitor renal function and urine elimination, hydration, fluid and electrolyte balance.

•Give ACE inhibitors to control hypertension and opioid analgesics for pain (Mallet, Lee, Mai, Lopez-Vargas, & Rangan, 2015).

•Provide fluids and foods based on the patient's condition, encourage increased fluids if the patient has a urinary tract infection, and restrict fluids if the patient has renal failure.

•Obtain specimens for urinalysis and culture and sensitivity as ordered to evaluate for hematuria, proteinuria, and infection; obtain specimens for laboratory tests for electrolyte levels (Reed-Gitomer, 2017).

•Allow the patient to verbalize his feelings and concerns, especially related to possible progression of the disease and renal failure; provide support and guidance.

•Prepare the patient for dialysis or renal replacement therapy if indicated.

•Encourage the parents of a child with the infantile form to obtain genetic counseling.

•Refer the patient and his family to community and social services.

CONCLUSION

Although there is no treatable cure for ADPKD, understanding the mode of inheritance and the implications is paramount. Patients with PKD having a basic understanding how the disease is inherited and establishing an accurate family history can appreciate how conditions are passed on in a family and passed down through generations. Managing the progression of the disease and the symptoms are incremental to slowing the growth of the cysts which may potentially lead to end stage renal disease (ESRD).

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